FEATURED CLINICAL CASE

Assessing the risk of sudden cardiac death

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Abstract

Detection of a major risk factor for sudden death in an otherwise asymptomatic person often raises major difficulties in management, particularly where the only treatment available is invasive, such as the implantable defibrillator. Recent guidelines have described the appropriate use of this technology, but often difficulty remains. This is particularly the case where the condition is newly recognised and its natural history not yet extensively described. A 63 year old man, whose condition was diagnosed as Brugada syndrome, in whom this problem is illustrated is described.

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A 63 year old man was referred for cardiological assessment in December 1999 with a history of intermittent flu like symptoms and abnormal ECGs. He had a history of recurrent episodes of pericarditis. He had no history of palpitations, dizziness, or syncope. Examination was unremarkable apart from an increased blood pressure of 170/90 mm Hg. Baseline investigations including haematology, biochemistry, and thyroid function tests were normal. A 12 lead ECG showed sinus bradycardia at 54 beats/min, first degree heart block (PR interval of 264 ms) and ST elevation in V1–V3 with a partial right bundle branch block pattern (fig 1). Review of notes showed similar ECG changes in 1987. His transthoracic echocardiogram showed normal left ventricular function and no regional wall motion abnormality. He underwent an exercise tolerance test, exercising for 6:13 minutes of the Bruce protocol with no symptoms or evidence of ischaemia.

The patient had a family history of sudden cardiac death. He had a sister who had died suddenly at the age of 55 years. A postmortem examination was not performed. A 12 lead ECG performed three years before her death in 1984 showed sinus rhythm, first degree heart block, and ST elevation in leads V1 and V2 (fig 2).

On the basis of ECG changes and the family history of sudden cardiac death, the patient's condition was diagnosed as Brugada syndrome.

Discussion

Brugada syndrome was first described in 1992.[1] This syndrome is diagnosed by characteristic ECG changes of right bundle branch block and ST segment elevation from V1–V3 in the context of sudden death (aborted or not) or syncope. Symptoms are attributable to either polymorphic ventricular tachycardia or ventricular fibrillation. It is important to recognise this characteristic ECG pattern as it is a marker for sudden cardiac death. The disease is responsible for up to 4–12% of unexpected sudden deaths and for up to 50% of sudden deaths in patients with a structurally normal

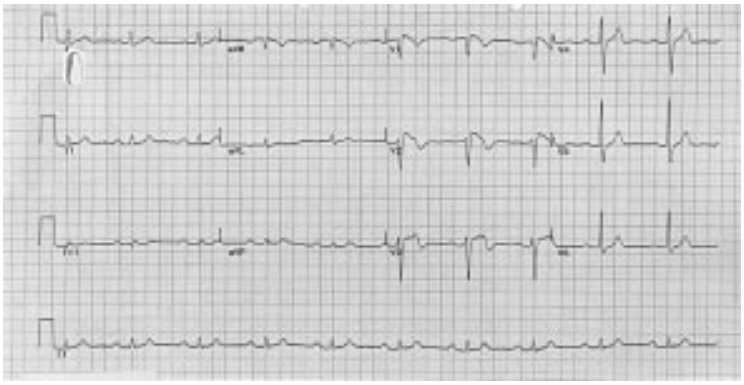

Figure 1 Twelve lead ECG showing sinus bradycardia, first degree atrioventricular block, incomplete right bundle branch block, and ST segment elevation in leads V1–V3.

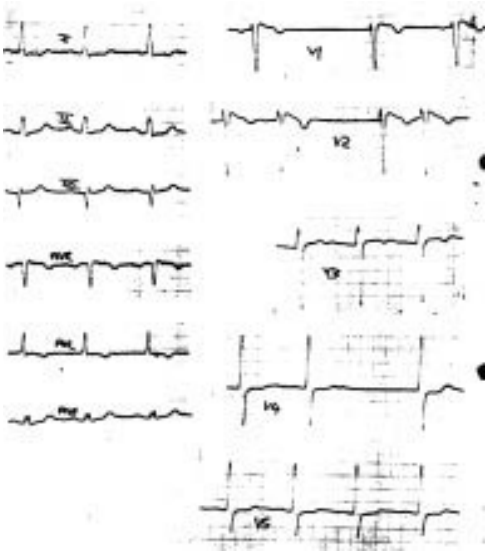

Figure 2 Twelve lead ECG showing sinus bradycardia, first degree atrioventricular block, incomplete right bundle branch block, and ST segment elevation in leads V1–V2.

heart.[2] The syndrome is estimated to have an autosomal dominant inheritance in 30% of families and no clear pattern of inheritance in 20% of families. The remaining 50% of cases are sporadic, suggesting a "de novo" mutation.[3] Mutations in the *SCN5A* gene encoding the cardiac sodium channel have been described but not all families with Brugada syndrome have a mutation of this gene, suggesting a heterogeneous genetic disease.[4]

Ailings and colleagues[4] reported that antiarrhythmic drug treatment with amiodarone, β blockers, or both did not prevent sudden cardiac death in Brugada syndrome and therefore current recommendations suggest use of an implantable cardioverter defibrillator in symptomatic patients.

However, the optimum management of asymptomatic patients with Brugada syndrome remains controversial, as the natural history of the disease in this group of patients is unclear. According to Brugada, asymptomatic patients recognised at random or discovered in a family study have a 27% incidence of arrhythmic events during a mean follow up of 34 months.[5] Patients with a family history of symptomatic Brugada syndrome, such as the patient in our case report, are reported to have an increased risk of sudden death.[6] This suggests an implantable cardioverter-defibrillator to be appropriate treatment. However, against this is a more recent study showing 0% cardiovascular mortality of asymptomatic patients with Brugada syndrome over a mean follow up period of 49 months.[7]

The dilemma we face is deciding on the appropriate management of our patient, who is 63 years old and asymptomatic but has a family history of sudden cardiac death. This is an interesting dilemma especially in view of a recent French study estimating a high prevalence (0.1%) of the typical ECG of the Brugada syndrome in an apparently healthy and asymptomatic French population.[7]

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Editor's comment

It is evident from this featured case that it was very difficult for the doctors looking after the patient to decide on the correct course of treatment. We would be very interested in receiving the views of readers on this interesting case report, as a rapid response. Such responses would be particular valuable if they refer to references to the current literature. I hope that we'll receive a large response from our readership.

ROGER HALL
Editor